Rhematologic Disorders in the Head and Neck

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Autoantibody relationships in connective tissue diseases

Disease	Autoantibody
Systemic lupus erythematosus	Anti-native DNA, Anti-Sm
Rheumatoid arthritis	Rheumatoid factor, Anti-RA33
Sjögren's syndrome	Anti-Ro(SS-A), Anti-La(SS-B)
Systemic sclerosis	Anti-Scl-70, Anti-centromere
Polymyositis/dermatomyositis	Anti-Jo-1
Mixed connective tissue disease	Anti-U1-RNP
Wegener's granulomatosis	c-ANCA

Histopathologic features

- Connective tissue and blood vessel inflammation
- fibrinoid deposits

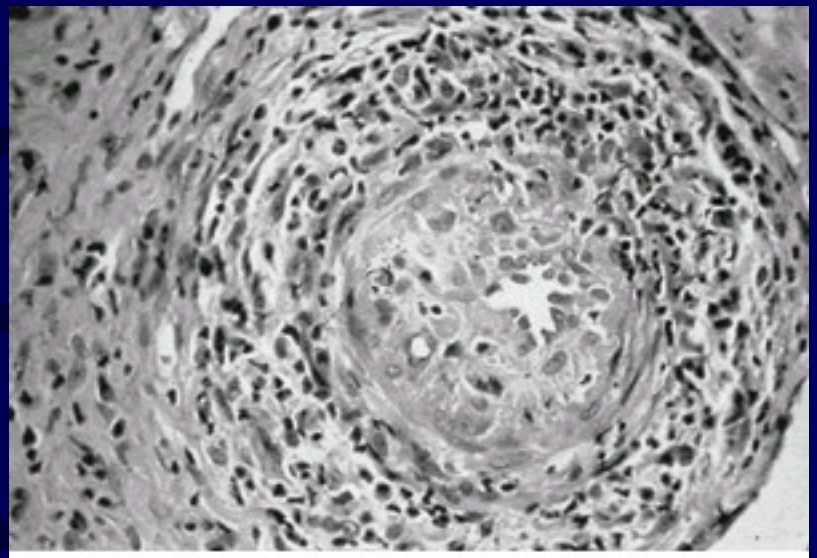

FIG. 16-1. Necrotizing and fibrinoid necrosis is seen in many connective tissue diseases. This biopsy from a patient with periarteritis nodosa shows an infiltration of eosinophils and polymorphonuclear leukocytes with medial necrosis and intimal proliferation.

Systemic Lupus Erythematosus

- Incidence of 1:2,000
- 9:1 ratio of women to men
- primarily affects women of childbearing age
- photosensitive skin eruptions
- nephritis
- pneumonitis
- myocarditis

Head & Neck Manifestations

- Malar “butterfly” rash in 64%
- Mucosal Ulcerations in 15%
- Nasal Septal Perforation in 3-5%
- Dysphagia in up to 25%
- TVC thickening or paralysis
- Cricoarytenoid arthritis
- Subglottic stenosis
- Enlargement of parotid gland 10%
- Xerostomia
- Cranial nerve palsies in 15%
- Autoimmune inner ear disease?

FIG. 16-2. Facial rash in the butterfly distribution typical of systemic lupus erythematosus.

Discoid Lupus

- Well-demarcated, erythematous, edematous papules
- depigment and scar on resolution
- Face is involved in 85% of cases
- Scalp involvement in 60% of cases
- Ear involved 44% of the time
- may have associated leukoplakia of tongue and oral mucosa

Treatment of SLE

- Avoidance of sun exposure
- NSAIDS
- topical and systemic steroids
- antimalarials
- low-dose methotrexate
- symptomatic treatment of lesions

Rheumatoid Arthritis

- Inflammation of synovial tissue with symmetric involvement of peripheral joints
- Affects 1% of the population
- 2-3 x more common in women
- juvenile form
- usu occurs in 40’s and 50’s

- TABLE 16-3. Diagnosis Rheumatoid arthritisa
- 1. Morning stiffness (³1 h)
- 2. Swelling of three or more joints
- 3. Swelling of hand joints (proximal interphalangeal,
- metacarpophalangeal, or wrist)
- 4. Symmetric joint swelling
- 5. Subcutaneous nodules
- 6. Serum rheumatoid factor
- 7. Radiographic evidence of erosions or
- periarticular osteopenia in hand or wrist joints
- aCriteria 1 to 4 must have been continuous for 6 weeks or longer and must be observed by a physician. A diagnosis of rheumatoid arthritis requires that four of the seven criteria be fulfilled.

Head & Neck Manifestations

- May affect the ossicles producing a conductive hearing loss
- Temporomandibular joint dysfunction
- Cricoarytenoid joint involvement in up to 86% of patients
- hoarseness in 30% of patients
- dyspnea on exertion
- anterior neck or ear pain
- globus sensation
- dysphagia
- ischemic recurrent nerve paralysis

Treatment

- Salicylates
- NSAIDS
- Gold Salts
- Penicillamine
- hydroxychloroquine
- immunosuppressive agents

Sjogren's Syndrome

- Immune-mediated destruction of exocrine glands
- sicca syndrome (lacrimal and salivary glands)
- Occurs in 1% of the general population
- Occurs in 10-15% of patients with rheumatoid arthritis
- 9:1 female to male distribution
- usu occurs between ages of 40 and 60.

- TABLE 16-4. Diagnosis Sjögren's syndromea,b
- 1. Dry eyes (>3 mo), sensation of sand or gravel in
- eyes, or use of tear substitutes >3 times a day
- 2. Dry mouth (>3 mo), recurrent or persistent
- swollen salivary glands, or frequent drinking of liquids to aid in swallowing dry foods
- 3. Schirmer-I test £5 mm in 5 min) or Rose bengal
- score ³4
- 4. >50 mononuclear cells/4 mm2 glandular tissue
- 5. Abnormal salivary scintigraphy or parotid
- sialography or unstimulated salivary flow £1.5
- mL in 15 min
- 6. Presence of anti-Ro/SS-A, anti-La/SS-B,
- antinuclear antibodies or rheumatoid factor
- aExclusion criteria: preexisting lymphoma, acquired immunodeficiency syndrome (AIDS), sarcoidosis, or graft-versus-host disease. bPresence of four or more criteria classifies primary Sjögren's syndrome with a sensitivity of 94% and a specificity of 94%.

Diagnosis

- Xerophthalmia and xerostomia
- minor salivary gland biopsy with heavy lymphocyte infiltration
- elevated RF and ANA
- Anti-Ro/SS-A in 60%
- Anti-La/SS-B in 30%

Treatment

- Increased oral fluid intake
- saliva substitutes
- pilocarpine
- nystatin for oral candidiasis
- close dental supervision

Systemic Sclerosis (Scleroderma)

- Sclerotic skin changes accompanied by multisystem disease
- increased deposition of collagen in the interstitium and intima of small arteries
- Raynaud’s phenomenon
- edema of the fingers and hands
- skin thickening
- arthralgias and muscle weakness
- Visceral involvement in GI tract, lung, heart, kidneys and thyroid

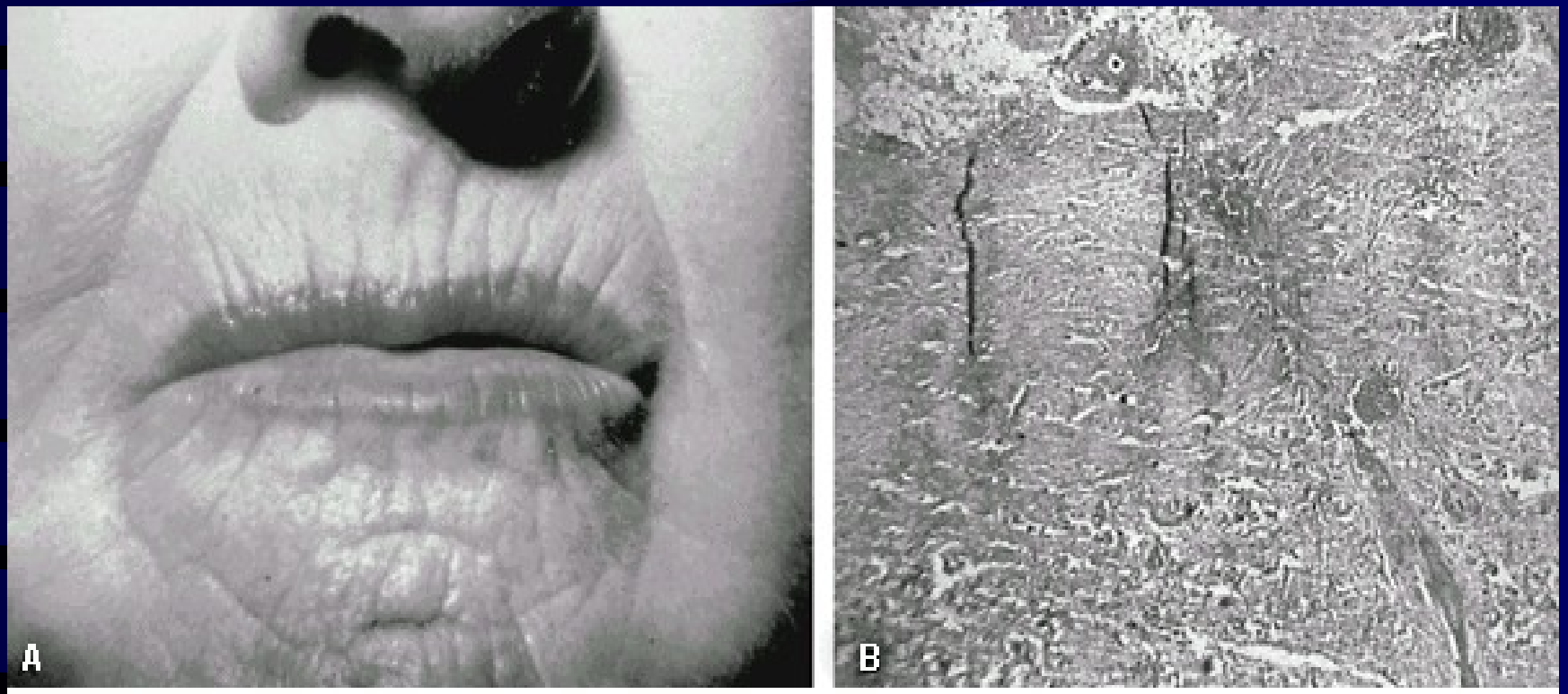

FIG. 16-3. A: The typical facies of scleroderma: tight skin, thin lips, and vertical perioral furrows. These external facial features are secondary to inflammatory foci in the dermis and subcutaneous tissues. B: Infiltrations of lymphocytes, plasma cells, and histiocytes.

Head and Neck Manifestations

- Tight skin, thin lips, vertical perioral furrows
- Dysphagia (decreased peristalsis)
- Decreased ability to open mouth
- telangiectasias in 19%
- calcinosis in 3%
- gingivitis and periodontal membrane thickening
- xerostomia or xerophthalmia in 25%
- voice change
- Raynaud’s phenomenon of tongue and trigeminal neuralgia

Treatment

- Calcium channel blockers for Raynaud’s
- H2 blockers for GERD
- NSAIDS for arthralgias and myalgias

Polymyositis and Dermatomyositis

- Proximal muscle weakness
- elevated serum skeletal muscle enzymes
- myopathic changes by electromyography
- muscle biopsy evidence of inflammation
- associated with SLE, scleroderma, RA
- assoc with a malignancy in 20% of cases

Head and Neck Manifestations

- Weakness of meck muscles in 50% of pts
- difficulty with phonation and deglutition (weak tongue)
- nasal regurgitation (weak palatal muscles)
- dysphagia in 30% of pts, due to weakness of upper esophagus, criocopharyngeus, pharynx and superior constrictors
- lesions of the eyelids, nose, cheeks
- stomatitis
- autoimmune inner ear disease

Treatment

- Steroids
- Methotrexate
- immunosuppressive agents
- H2 blockers
- metoclopramide

Relapsing Polychondritis

- Episodic recurring inflammation of cartilaginous structures and replacement by granulation tissue and fibrosis
- More common in women than in men
- Onset between ages 35 and 45

Diagnostic Criteria

- Recurrent chondritis of auricles
- nonerosive inflammatory polyarthritis
- chondritis of the nasal cartilages
- inflammation of ocular structures
- chondritis of laryngeal or tracheal cartilages
- chochlear or vestibular damage

Head and Neck Manifestations

- Auricular chondritis in 90%. Sudden onset of erythema and pain, sparing the EAC and resolving in 5-10 days. Patients may have OM or SNHL. 49% will have inner ear symptoms.
- Chondritis of nasal cartilages in 75%
- Laryngeal involvement presenting with a non-productive cough, hoarseness, and stridor. Upper respiratory tract is involved in 53% of patients

Treatment

- Salicylates and NSAIDS
- Steroids for life-threatening manifestations
- dapsone (reduces lysozymes)

Mixed Connective Tissue Disease

- Coexisting features of SLE, systemic sclerosis, polymyositis, and dermatomyositis
- high titers of anti-U1 RNP (ribonucleoprotein)
- prevalence is unknown
- 80% of patients are women

Diagnostic Criteria

- Elevated anti-U1 RNP + 3 of the following:
- hand edema
- synovitis
- myositis
- Raynaud’s phenomenon
- acrosclerosis
- (pulmonary involvement is common)

Head and Neck Manifestations

- Malar rash
- discoid lupus
- sclerodermatous skin thickening
- oral mucosal ulceration
- nasal septal perforation
- Sicca complex
- esophageal dysfunction

Polyarteritis Nodosa

- Affects males 2-3 x more than women
- onset is 50’s to 60’s
- Hepatitis B antigen in 30% of patients
- vasculitis of small and medium sized arteries
- involves GI tract hepatobiliary system, kidneys, pancreas and skeletal muscles.
- Rare bilateral sudden sensorineural hearing loss, thought to be due to thromboembolic occlusion of end arteries.
- Ulceration of nasal, buccal and soft palate mucosa

Churg-Strauss Syndrome

- “allergic angiitis granulomatosis”
- variant of polyarteritis nodosa with systemic vasculitis, asthma, tissue eosinophilia
- lungs are always involved
- 70% of patients with nasal obstruction and rhinorrhea

Wegener's Granulomatosis

- Respiratory tract granulomas, vasculitis, glomerulonephritis
- bilateral pneumonitis in 95% of patients
- chronic sinusitis in 90%
- mucosal ulceration of nasopharynx in 75%
- evidence of renal disease in 80%
- nasal crusting, epistaxis, and rhinorrhea
- hyperplasia of gingiva and gingivitis
- edema and ulceration of larynx in 25% (subglottic stenosis in 8.5%)
- serous otitis media in 25%

- Necrotizing granulomatous vasculitis
- cytoplasmic staining antineutrophil cytoplasmic antibody
  (c-ANCA)

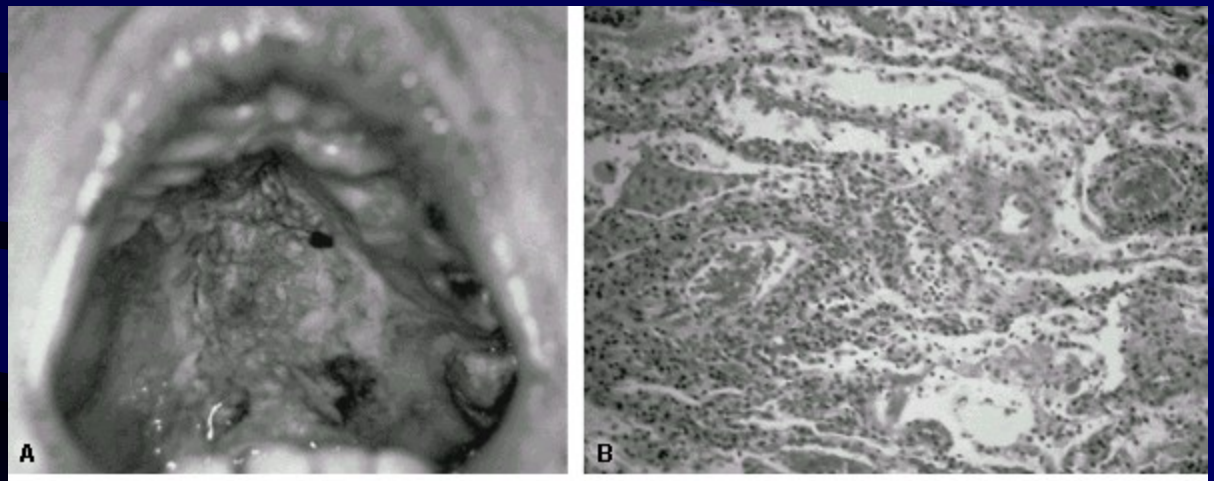

FIG. 16-4. A: Wegener's granulomatosis involving the palate. B: This lung biopsy from a patient with Wegener's granulomatosis demonstrates the typical vasculitis and necrotizing tissue reaction.

Giant Cell Arteritis

- Focal granulomatous inflammation of medium and small arteries
- Headache is initial symptom in 47%
- Headaches occur in 90%
- Temporal arteries involved only 50% of the time
- tender scalp and jaw claudication in 50%
- vertigo and hearing loss, rarely
- dysphagia (involvement of ascending pharyngeal artery)
- cranial nerve deficits (intracranial disease)

-ESR usually > 50 mm/hr
-diagnosed with temporal artery biopsy

FIG. 16-5. Giant cell arteritis typically affects arteries with an elastic lamina. This temporal artery biopsy demonstrates the characteristic polymorphonuclear and mononuclear infiltration, medial necrosis, disruption of the elastic lamina, and giant cell formation.

Polyarteritis Rheumatica

- Muscular pain
- morning stiffness in proximal muscles
- elevated ESR without inflammatory joint or muscle disease
- occurs in 50% of patients with giant cell arteritis
- may begin with fever, weight loss, malaise

Behcet's Disease

- Oral and genital ulcers (punched out ulcers with surrounding erythema, covered by a pseudomembrane)
- uveitis or iritis
- progressive SNHL, tinnitis, vertigo
- may lead to ulceration of nasal, laryngeal, tracheal mucosa

Cogan’s Syndrome

- Audiovestibular dysfunction (fluctuating hearing loss, vertigo, tinnitus, aural pressure). Symptoms disappear and reappear months later.
- interstitial keratitis
- nonreactive tests for syphilis
- steroid responsive

Kawasaki Disease

- “mucocutaneous lymph node syndrome”
- pediatric age group
- fever, conjunctivitis, red and dry lips, erythema of the oral mucosa, polymorphous truncal rash, desquamation of the fingers and toes, cervical lymphadenopathy.
- 1-2% mortality due to cardiac abnormalities